Torsion of a wandering spleen with preserved vascular supply

Torsión de un bazo migratorio con aporte vascular conservado doi: 10.61997/bjm.v14i3.484

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ABSTRACT

Introduction: Wandering spleen is a rare clinical entity caused by congenital or acquired laxity, elongation, or absence of the splenic ligaments, allowing the spleen to migrate from its usual anatomical position. The most serious complication is torsion of the splenic vascular pedicle, which can lead to infarction, peritonitis, and, if not promptly treated, death. **Case report:** We report the case of a 2-year-old female with a history of intermittent abdominal pain and a recent finding of a mobile infraumbilical mass. She was referred with a presumptive diagnosis of an abdominal tumor. Abdominal ultrasound revealed an ectopic spleen with torsion of the vascular pedicle. Despite the torsion, splenic vascularization was preserved. Emergency laparotomy was performed. Intraoperatively, a 720-degree torsion of the splenic pedicle was identified. Manual detorsion was performed, and splenopexy was achieved. The postoperative course was uneventful. **Conclusions:** This case underscores the importance of early diagnosis and prompt surgical intervention in patients with torsion of a wandering spleen. Ultrasound with Doppler imaging is an effective first-line diagnostic tool. When the spleen is viable, splenopexy remains the treatment of choice in pediatric patients.

Keywords: Wandering spleen; Splenic torsion; Splenopexy; Ectopic spleen

RESUMEN

Introducción: El bazo migratorio es una entidad clínica rara causada por la laxitud, elongación o ausencia congénita o adquirida de los ligamentos esplénicos, lo que permite que el bazo se desplace de su posición anatómica habitual. La complicación más grave es la torsión del pedículo vascular esplénico, que puede llevar al infarto, peritonitis y, si no se trata oportunamente, a la muerte. **Presentación del caso:** Se presenta el caso de una paciente de 2 años con antecedentes de dolor abdominal intermitente y hallazgo reciente de una masa infraumbilical móvil. Fue derivada con diagnóstico presuntivo de tumor abdominal. La ecografía abdominal evidenció un bazo ectópico con torsión del pedículo vascular. A pesar de la torsión, la vascularización esplénica se encontraba conservada. Se realizó laparotomía de urgencia donde se constató una torsión de 720 grados del pedículo esplénico. Se practicó destorsión manual y esplenopexia. La evolución postoperatoria fue favorable. **Conclusiones:** Este caso resalta la importancia del diagnóstico precoz y la intervención quirúrgica oportuna en pacientes con torsión de bazo migratorio. La ecografía Doppler es una herramienta diagnóstica efectiva de primera línea. Cuando el bazo es viable, la esplenopexia continúa siendo el tratamiento de elección en pacientes pediátricos

Palabras claves: Bazo errante; Torsión esplénica; Esplenopexia; Bazo ectópico

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INTRODUCTION

First described as an anatomical curiosity in the 17th century, the wandering spleen has evolved from a postmortem finding to a recognized surgical emergency, particularly in pediatric patients when complicated by torsion.1

The word spleen derives from the Greek splen and the Latin lien. It is the second largest organ of the reticuloendothelial system, with a dark reddish color, and its size varies depending on age. It is in the left hypochondrium, positioned between the gastric fundus and the diaphragm, and is partially protected by the rib cage (between the 9th and 11th ribs). The spleen is anchored in its anatomical position by several suspensory ligaments, including the splenophrenic, splenocolic, splenopancreatic, gastrosplenic, and splenorenal ligaments, which maintain it fixed within the splenic fossa.^{2,3} Wandering spleen is characterized by abnormal positioning of the spleen within the abdominal cavity due to laxity, elongation, or congenital absence of the splenic suspensory ligaments. These abnormalities predispose to elongation of the vascular pedicle, facilitating partial or complete torsion of the splenic axis.2

Although its incidence in children is low, the condition carries a high risk of acute abdominal complications, including infarction and peritonitis. In younger patients, clinical presentation is often nonspecific and overlaps with more common causes of abdominal pain, such as gastroenteritis, appendicitis, or intestinal parasitism, making preoperative diagnosis particularly challenging.^{2,3}

Here, we report the case of a 2-year-old girl with torsion of a wandering spleen, in whom diagnosis was established through imaging. The case highlights the critical role of ultrasound, and the importance of timely surgical intervention aimed at spleen preservation whenever feasible.

CASE REPORT

A 2-year-old female patient with a history of recurrent abdominal pain over several months, previously treated twice for intestinal parasitosis (Giardia lamblia), was brought to a primary healthcare center after her mother noticed a mobile, non-tender infraumbilical mass during bathing. Based on this finding, she was urgently referred to the Pediatric Hospital of Camagüey, Cuba, with a presumptive diagnosis of an abdominal tumor.

During her evaluation in the Pediatric Surgery Department, the mother reported that the child had a generally poor appetite but remained active. Since the age of one, she had experienced intermittent episodes of abdominal pain, sudden in onset, unrelated to physical activity, and often resolving spontaneously without medication. At the time of examination, the child had been asymptomatic for over two weeks, but the mother had noticed infraumbilical swelling earlier that morning. There were no associated symptoms such as fever, vomiting, diarrhea, asthenia, anorexia, or weight loss.

On physical examination, the abdomen was flat with normal respiratory movements. There was visible infraumbilical swelling without skin discoloration. A palpable, mobile, smooth-surfaced, non-tender mass measuring approximately 6 cm was noted in the hypogastric region. The bowel sounds were normal. The remainder of the physical examination was unremarkable.

Laboratory investigations revealed a white blood cell count of 9.3×10^3 /mm³, with neutrophils at 61%, lymphocytes 38%, and eosinophils 1%. Hematocrit was 32%, hemoglobin 105.6 g/l, platelets 300,000/mm³, and total protein 58 g/l.

Abdominal ultrasonography identified the spleen located in the mesogastrium, measuring 65 × 53 × 38 mm. The organ appeared smooth, mobile, and non-tender. Apparent splenic hilar lymphadenopathy was noted, with the largest node measuring 13 × 8 mm. No other abnormalities were found. Color Doppler imaging demonstrated torsion of the vascular pedicle; however, splenic blood flow was preserved.

An emergency abdominal computed tomography (CT) scan was deemed unnecessary, as the ultrasonographic findings were conclusive. Performing a CT scan would have resulted only in a delay of the urgently indicated surgical intervention.

Diagnosis and Management

A diagnosis of subacute splenic torsion was established, and the patient was admitted for urgent surgical management. Thirty minutes prior to surgery, the first dose of prophylactic cefazolin was administered.

Surgical Procedure

Given the team's limited experience with laparoscopic approaches for this procedure in a 2-year-old patient, an open approach was selected. A left subcostal incision measuring approximately 5 cm was made, 2 cm below the left costal margin. Dissection proceeded through anatomical layers until the peritoneal cavity was entered. Upon entry, moderate amount of serous fluid was found and aspirated.

The spleen was identified, gently mobilized toward the surgical field and successfully exteriorized. Notably, there was a complete absence of splenic ligamentous attachments. The splenic pedicle was found to be twisted to 720 degrees; however, the spleen maintained a normal color, with a smooth surface and no evidence of recent or previous ischemia or necrosis. (Figure 1).



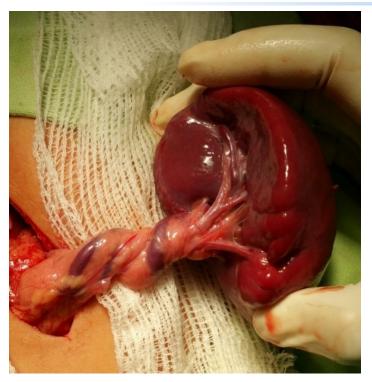


Figure 1. Exteriorized spleen with torsion of its vascular pedicle.

Manual untwisting of the splenic pedicle was performed. The vascular pedicle appeared thickened, with tortuous vessels of greater diameter than expected for the patient's age. Multiple lymph nodes were observed along the pedicle. (Figure 2).

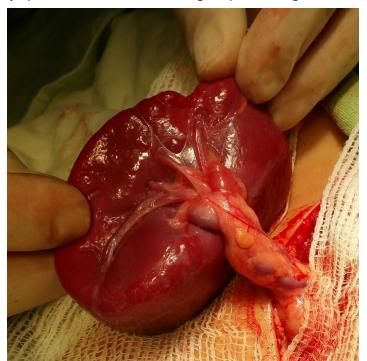


Figure 2. Spleen after manual untwisting of the vascular pedicle.

Given the spleen preserved its vascular supply, a splenic pocket was created using the omentum. The omental flap was

anchored to the splenic capsule using 4-0 Vicryl sutures. The spleen was then fixed to the left lateral abdominal wall in the left hypochondrium using four 3-0 Vicryl sutures (Figures 3 and 4). The abdominal wall was closed in anatomical layers with 3-0 Vicryl, 4-0 Vicryl, and 4-0 Monocryl sutures.

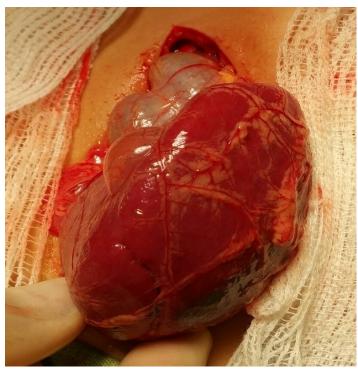


Figure 3. Omental pocket covering the spleen.



Figure 4. Suture attachments to the abdominal wall.

Postoperative Course

The postoperative recovery was uneventful. The patient was discharged 48 hours after surgery in good general condition. At the six-month follow-up, abdominal ultrasonography confirmed the spleen's position in the left hypochondrium. It measured 63 × 58 × 42 mm, with homogeneous echotexture and smooth surface (Figure 5). Based on these findings, the patient was discharged from pediatric surgical follow-up.



Figure 5. Postoperative ultrasonography of the spleen.

DISCUSSION

The first reported case of a wandering spleen in the medical literature is attributed to Dr. Józef Dietl, a Polish physician, who published the case in 1854 in the Diary of the Warsaw Medical Society and the Wiener Medizinische Wochenschrift. Two years later, he reported a second case, diagnosed postmortem, in the same journal,4 and, in 1863 he described a third patient with this condition in a medical journal he had founded himself.⁵ In this publication, he noted that the pathology could be fatal, as its complications could lead to peritonitis and ultimately death. Dr. Dietl was among the first physicians to propose that the condition was not linked to an individual's temperament, but rather to ligamentous laxity, elongation, or hypoplasia, which allowed the spleen to migrate.1

Wandering spleen is most frequently found in children and women of childbearing age, with fewer than 500 cases reported in the general medical literature. The incidence in children is even lower, reported in around 0.2%.^{2,6} As Dr. Dietl proposed, this condition is characterized by the spleen being at any part of the abdominal cavity. 1 Its occurrence

is considered multifactorial, as its origin may be either congenital or acquired.^{2,3,7}

Congenital wandering spleen typically results from the failure of fusion between the dorsal mesogastrium and the posterior peritoneum during the fifth to sixth weeks of embryonic development. This developmental defect leads to the absence, malformation, or laxity of one or more splenic ligaments structures responsible for maintaining the spleen in its anatomical position within the left upper quadrant. As a result, the spleen may become hypermobile, predisposing the vascular pedicle to torsion.^{2,6,8}

Acquired forms of wandering spleen, although less common in the pediatric population, have been reported and are associated with factors such as abdominal trauma, connective tissue disorders, multiparity, renal agenesis, and hormonal changes during pregnancy. These conditions may either damage the ligamentous supports or contribute to their hyperlaxity, facilitating abnormal splenic mobility. Notably, acquired wandering spleen in pediatric patients has been documented in association with systemic conditions such as immunoglobulin deficiency, infectious mononucleosis, malaria, Gaucher disease, Hodgkin lymphoma, and DiGeorge syndrome.^{2,3} These associations highlight the diverse etiological spectrum of wandering spleen in younger patients. In our case, the complete absence of all splenic ligamentous attachments strongly supports a congenital origin. This observation aligns with findings from several pediatric wandering spleen reviews, which emphasize that toddlers rarely present with any relevant clinical antecedents.2 Consequently, the younger the patient, the more likely the etiology is congenital rather than acquired.

The diagnosis of wandering spleen is often challenging due to its nonspecific clinical manifestations. Asymptomatic cases can be undiagnosed or incidentally discovered. Symptomatic patients, however, may present with a wide spectrum of clinical signs depending on the degree and nature of splenic torsion, the presence of accompanying symptoms, and whether other intra-abdominal organs are involved in the torsion process.6,7

The intensity and pattern of the pain are generally proportional to the degree of torsion. Mild torsion may cause chronic abdominal pain due to venous congestion; moderate torsion often manifests as intermittent pain caused by recurrent torsion and detorsion of the splenic pedicle; and severe torsion almost invariably leads to splenic infarction or rupture. In such cases, signs of peritonitis may appear, and patients may progress to hypovolemic shock due to hemoperitoneum, the most serious clinical scenario.^{2,6}

Accompanying symptoms such as nausea, anorexia, low-



grade fever, and fatigue may also be present,2,7,8 and can delay diagnosis by mimicking more common pediatric gastrointestinal conditions. The clinical course becomes more complex when other intra-abdominal organs are involved. The stomach, pancreas, or colon may rotate along with the spleen due to shared ligamentous attachments or close anatomical relationships.9 This can result in gastric volvulus, gastric outlet obstruction (even rare), pancreatic torsion, or colonic obstruction. These complications significantly increase morbidity and may present additional symptoms such as hematemesis, back pain, or even signs of pancreatitis or bowel ischemia. 10,11 Although rare, gastric varices or variceal bleeding may occur in patients with splenic torsion and can be further aggravated by coexisting gastric volvulus, contributing to a clinical picture of left-sided portal hypertension. 4,11

Interestingly, our patient presented with a non-tender abdominal mass, preceded by a history of recurrent abdominal pain and two previous treatments for suspected intestinal parasitism. This presentation is consistent with findings from a study by Verma et al, 12 which highlights that recurrent abdominal pain is a frequent complaint during childhood and is often initially attributed to gastroenterocolitis or parasitic infections.

Preoperative diagnosis remains particularly challenging due to the nonspecific nature of the symptoms and the condition's low incidence. 8,9,11 Nonetheless, abdominal ultrasound with Doppler has proven to be the most practical and effective initial imaging modality in children. It can reveal the absence of the spleen from its usual anatomical position and locate it elsewhere within the abdominal cavity. However, ultrasound may be limited by bowel gas, difficulty in evaluating adjacent viscera, and challenges in identifying the twisted pedicle or infarcted spleen. CT scanning is considered the imaging modality of choice for confirming wandering spleen, as it can clearly demonstrate the ectopic spleen, identify the "whirl sign" of twisted splenic vessels, and detect associated findings such as ascites or involvement of neighboring organs. While scintigraphy and angiography may be used for diagnosis, their invasive nature and higher costs make them less favorable in routine pediatric evaluation. 6,13,14

Although it is acknowledged that abdominal ultrasound is an operator-dependent study, in our case, it enabled accurate and timely identification of both the ectopic spleen and the twisted vascular pedicle, while also confirming preserved splenic perfusion on ecodoppler. This facilitated prompt surgical management and prevented delays that might have occurred from ordering a CT scan.

The management of wandering spleen has evolved significantly since the first successful splenectomy was performed by Dr. Martin in 1877 on a 31-year-old woman with a migratory spleen, in his words: "This, gentlemen, is the case I have to report to you. It proves undoubtedly that we can exist without the spleen, that splenotomy does not change the constitution of the blood", a statement published in the British Medical Journal in 1878.15

While this historic milestone demonstrated the feasibility of life without a spleen, current understanding emphasizes the importance of splenic preservation, when possible, due to the immunologic spleen function overall to prevent post-splenectomy sepsis, particularly from encapsulated pathogens such as Streptococcus pneumoniae, Neisseria meningitidis, and Hemophilus influenzae.^{2,8,16,17}

As conservative management in asymptomatic patients has been associated with a complication rate as high as 65%,9 surgery remains the gold standard for the treatment of WS. The two primary surgical techniques are splenopexy and splenectomy, via open or laparoscopic surgery, with the choice depending on spleen viability, size, and associated complications such as hypersplenism or thrombosis. In cases of infarction, rupture, or thrombosis of the splenic or portal veins, splenectomy becomes necessary. In these cases, vaccinations for encapsulated organisms should be given after surgery. 2,16,17

Surgical techniques have progressed from the traditional Bardenheuer's procedure, where the spleen is secured in a retroperitoneal pouch, 8,18 to modern laparoscopic splenopexy, now considered the gold standard in suitable cases. 2,17-20

Recent reviews continue to emphasize organ sparing and minimally invasive techniques for wandering spleen in children. 17-19 In one of the largest paediatric syntheses to date, Ganarin A, et al 20 analysed 197 cases and found that splenectomy was performed in 54.8% of patients, whereas splenopexy accounted for 39%; notably, almost half of the splenopexies were completed laparoscopically, predominantly with mesh (45.5%) or a retroperitoneal pouch (30.9%), and achieved a 94.8% success rate. In contrast, the single centre series reported by Wang Z, et al 9 underscores the challenges of acute torsion: all six children treated between 2016 and 2021 required emergent splenectomy because of complete or near complete infarction, despite timely imaging and laparoscopic access. Together, these findings illustrate that while elective or sub acute presentations increasingly benefit from laparoscopic splenopexy, irreversible vascular compromise still mandates splenectomy in a significant subset of cases.

In our case, despite two complete torsions of the vascular pedicle, splenic perfusion was preserved. Therefore, manual detorsion, creation of a splenic pocket with omentum, and splenopexy was the treatment of choice.



CONCLUSIONS

Wandering spleen is a rare but serious condition in children that can lead to life-threatening complications if not promptly diagnosed and treated. Early recognition, based on clinical suspicion and supported by ultrasound or CT, is essential. When the spleen is viable, splenopexy should be the preferred treatment to preserve its immunologic function. Despite minimally invasive approaches increasingly favored in pediatric patients with excellent outcomes, this procedure must be carried out by surgeons with specialized training in the technique and access to proper surgical equipment.

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Disclosure

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